



Conn's Syndrome Masquerading as Hypokalemic Paralysis

Authors

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Abstract

Hypertension is a globally prevalent disease with significant morbidity and mortality. While the usual presentation is non-specific, or related to end-organ symptoms like dizziness, headache, encephalopathy, cerebrovascular accident, palpitations, acute pulmonary odema, aortic dissection or renal failure, we report an atypical presentation where the patient presents to the Emergency Department with hypokalemia related weakness, and step-wise sequential evaluation lead to the detection of Secondary Hypertension, and Primary Aldosteronism due to a left Adrenal Adenoma.

Case Report

A 44 year old Female presented in the Medicine Emergency Department with complaints of sudden, symmetrical and progressive weakness of both lower limbs of 12-14 hours duration. However, the weakness had not involved the trunk or upper limb muscles. She denied any bladder, bowel, sensory or higher function complaints. There was no history of recent fever, cough, loose stools, vaccinations, indigenous or allopathic medications, prescribed or otherwise. There was history of occasional incidental high blood pressure readings, but never medicated. At the time of first evaluation, she was found to be moderately built, normal body habitus, BMI 22, blood pressure was 150/80 mm Hg; had a diffusely enlarged thyroid gland, normal pulses, no oedema, or hyperpigmentation. Systemic examinations of Cardiovascular, GIT and Respiratory tract were within normal limits. Neurological evaluation revealed flaccid weakness of both lower limbs with power 2/5, upper limb 4/5, absent deep tendon reflexes,

bilateral Flexor Plantar reflexes, intact Sensory and Cranial Nerve Examination and spared bladder and bowel function. Her personal, family, dietary menstrual and gestational history was unremarkable.

During the course of hospitalization, initial Haematology and Biochemistry reports were within normal limits, with the exception of Potassium values (K-1.8 meq/l). The ECG findings were consistent with hypokalemia. ESR-30. In view of the hypertension and hypokalemia, workup for secondary causes of hypertension was begun. Viral Markers (HIV, HbS Ag, HCV) were non-reactive, ANA Screening was Negative. The Goitre was evaluated; TSH- 3.2, USG Neck revealed diffusely enlarged thyroid gland with possibly colloid nodules. Renal artery Doppler failed to reveal any significant Main Renal Artery stenosis. USG Abdomen revealed only a small anterior myometrial fibroid. Meanwhile, Potassium Supplementation had improved the muscle weakness. Arterial blood gas analysis (ABG) revealed pH -7.46, HCO₃ - 32, CO₂ - 37.8

mm Hg. i.e metabolic alkalosis. Normal Cortisol levels in morning blood samples ruled out Cushing's syndrome. Urine 24 hour samples for estimation of Vinylmandelic Acid (VMA), Metanephrines, and Nor-metanephrines were within normal limits.

Despite a normal USG Abdomen, the coincidental hypokalemia and hypertension had raised the probability of Conn's syndrome. Hence, subsequent to normalization of Potassium levels, Serum aldosterone and plasma renin activity was measured in supine position. Serum Aldosterone Concentration (Plasma Aldosterone Concentration; PAC) was 45.40 (N 1.76-23.2), Direct Renin 0.884uIU/ml (N 2.8-39.9). The Aldosterone/Renin Ratio (ARR) was 54.177 ng/dL/uIU/ml. This strongly favoured Hyperaldosteronism, hence CECT of abdomen was done. The CECT Abdomen revealed "a Well Defined Left Adrenal Lesion, in the body of the gland, measuring 1.1x 1.1 cm, with central fat attenuation (4 HU), with heterogenous moderate enhancement in the arterial phase , rapid wash out in subsequent phases- possibly an Adrenal Adenoma ". The patient was given the option of surgical intervention. She has opted for conservative management with Amlodipine, Aldactone and losartan. She is presently asymptomatic, and is being kept in regular follow-up. Subsequent Potassium and Blood pressure readings were within normal limits. A follow through imaging is being planned after a year, to assess Adenoma growth.

Review of Literature

Hypertension has been now recognized as one of the leading causes of the global burden of Disease, accounting for an average of at least 7.6 million deaths and 92 million disability-adjusted life years worldwide as of 2001. The spectrum of environmental and genetic factors contributing to regional and racial variations in hypertension prevalence, and its etio-pathogenesis is still evolving. While Primary hypertension remains the most common cause, In about 5–20% of

hypertensive patients, a specific underlying disorder causing the elevation of blood pressure can be identified –hence, called as "Secondary Hypertension". Endocrine causes of Secondary Hypertension are now being increasingly recognized. However, physician mindset and lack of accessibility to specialized tests has contributed to undermining of the true incidence and prevalence of Endocrine Hypertension.

Acute onset Neuro- Muscular weakness in the tropical humid coastal climate of Kerala has multiple etiologies. An uncommon, but easily recognized and promptly responsive cause is Acute hypokalemic paralysis .Hypokalemia has varied clinical presentation, ranging from mild myalgia and lower limb weakness to fatal respiratory paralysis and arrhythmias. Reduced intake, Renal and non-renal losses, endocrine dysfunction and some inherited conditions like channelopathies might cause Hypokalemia.

Our patient had presented with hypokalemia associated weakness at the age of 44, and detected to have hypertension. An inconspicuous past, lack of Family history, normal renal, thyroid and LFT and absence of any drug history pointed towards a secondary Endocrine cause of hypokalemia. The ABG finding of metabolic alkalosis pointed to disease like Hyperaldosteronism -Conn's syndrome, Inherited conditions like Bartter, Gitelman and Liddle syndrome, Syndrome of Apparent Mineralocorticoid excess, Glucocorticoid Remediable Aldosteronism, and Diuretic use. The later age of onset, low Renin levels, and absence of polyuria ruled out the Autosomal recessive Gitelman's syndrome. Presence of hypertension and Hypo-Reninemic Hyperaldosteronism ruled out Bartter's syndrome. The triad of Hypokalemia, Hypertension and Metabolic Alkalosis is common in hyperaldosteronism. The elevated Aldosterone Renin Ratio confirmed Primary Hyperaldosteronism, and CECT abdomen confirmed the diagnosis of Adrenal Adenoma.

Conn's Syndrome or Primary Aldosteronism was first described by Conn in 1955. Secondary Hypertension is being now increasingly diagnosed

secondary to Conn's Syndrome. Aldosterone regulates renal fluid and sodium retention in lieu of Potassium excretion. Hyperaldosteronism thus causes Hypertension, Hypervolemia, suppressed Renin secretion and Hypokalemia. Solitary Aldosterone secreting Adenoma is the most common cause of Primary Aldosteronism, accounting for 65% cases. Other causes like Bilateral hyperplasia aldosteronism (BHA), Adrenocortical carcinoma, and Glucocorticoid-remediable aldosteronism (GRA) are relatively less frequent.

The most common presentation of Adrenal Adenoma is as Hypertension, which might be refractory, easy propensity to develop Hypokalemia with or without Diuretic use and Metabolic Alkalosis in a patient of 30-60 years age. Incidence is more in females. The Adenoma has been described as a small, less than 2 mm nodule, most frequently in left Adrenal, in middle age females with more profound hypokalemia, hypertension and elevated Aldosterone levels. The classical Triad of Hypokalemia, elevated Aldosterone and suppressed Renin is not mandatory to diagnose Primary Aldosteronism, as Hypokalemia might be seen only in 9-37% cases. PA should be suspected in the following cohort of patients: 1) Hypertension with hypokalemia 2)Resistant Hypertension (i.e uncontrolled with three anti-hypertensives, including diuretics) 3) Severe hypertension (>160/100mm Hg) 4) Hypertension with Adrenal Incidentaloma 5) Young onset HTN, under age 20.

While both CT and MRI can be used for imaging adrenal lesions, CECT Abdomen is capable of detecting Adrenal adenoma or hyperplasia with a sensitivity of atleast 85%. Percutaneous Transfemoral Bilateral Adrenal Venous sampling can localise the side with increased Aldosterone secretion, when CT scan is ambiguous. Treatment options include medical management with salt restriction and Aldosterone antagonist. The treatment of choice however is laparoscopic excision of the tumour. Review of literature showed prompt response of hypokalemia to

intervention, but hypertension tended to be persistent, especially in those with long standing disease duration, elderly and renal impairment. In our case, the patient expressed her clear unwillingness to undergo any surgical intervention, and hence was initiated on Aldosterone antagonist and anti-hypertensives. At present, her blood pressure and Serum Potassium Levels are well controlled.

Conclusion

Acute onset areflexic neuromuscular weakness is a very common presentation in the Emergency Dept of the tropical weather of South India. It is essential to look for hypokalemia and hypertension in such cases. A high clinical index of suspicion and sequential approach in such scenario might unmask causes other than Guillain Barre Syndrome, Myasthnia Gravis, and Hypokalemic periodic Paralysis. Conn's Syndrome is a potentially treatable condition, which if treated appropriately can prevent deleterious end organ dysfunction.

Disclosure

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